Chapter 31

Common Neurological Problems

Pediatric Neurological Examination

- **Mental status**
  - Is the child alert? Is the child appropriately oriented (based on age)?
  - Is the child age-appropriately interactive? If upset, is the child consolable?
- **Cranial nerves** (modified for the young child)
  - Cranial nerve (CN) II: visual acuity. In a young child, it is unlikely that the optic nerve can be visualized; however, several methods can be used to determine a young child's visual acuity
    - Does the child track objects (eg, a name badge, face, or small toy)?
    - How small an object will the child reach for?
    - Visual fields: if there is a small toy in front of the child, does the child look when another one is brought out from behind the examiner's back?
  - CN III, IV, VI: extraocular muscles, pupils, and ptosis
    - Use an object to check tracking, or spin the patient around in different directions to see if the patient's eyes move appropriately
  - CN V: facial sensation, muscles of mastication, corneal reflex
    - Does the patient respond to touch on the face?
    - If concerned, test corneal reflexes
  - CN VII: facial muscle strength and symmetry
    - Observe the patient crying, smiling, etc; compare sides
  - CN VIII: hearing
    - For older children, whisper a question in each ear and determine if the child can answer
    - For infants, clap hands or ring a bell and watch for a blink or other response

- CN IX, X, XII: palate elevation, straight tongue protrusion, strength with tongue deviation laterally
  - Observe swallowing and speech
  - Observe patient crying; observe palate elevation and tongue movements
- CN XI: sternocleidomastoid and trapezius strength
  - Have patient rotate head against resistance in each direction
  - Have patient shrug shoulders against resistance

- Coordination
  - Have child perform finger and foot tap (rapid alternating movements); observe movements for accuracy and rhythm
  - Have child bring a finger to the nose and heel to the shin; observe for ataxia or inability to hit the mark
  - Note abnormal movements
  - Modifications
    - Watch patient reach for and manipulate objects
    - If the child is old enough, ask the child to touch different parts of a toy
- Motor
  - Strength
    - Infants are best examined when they are fighting the examination or crying; does the fighting infant move all extremities? If so, observe for any differences (eg, can the infant push the examiner away equally with bilateral upper and lower extremities?)
    - Examine older children in the same manner as adolescents and adults
  - Tone
    - Patient needs to be awake and calm
    - For infants: test with patient's head midline (reflexes can be induced and examination findings changed if the head is turned)
      - Hypotonia (see Evaluating for Hypotonia)
      - Hypertonia
        - Note resistance to passive motion
        - Vertical suspension: legs crossing or "scissoring" suggests hypertonia
    - For older children: passively move the patient at variable

rates; note increases or decreases in tone, "catches," or rigidity

- Reflexes
  - Crossed adductors can be normal in children up to 12 months old
  - Babinski reflex/extensor plantar response (the big toe turns upward and other toes fan out when foot is tickled): can be normal up to 2½ years old
  - Unsustained clonus can be common in a healthy neonate
  - Reflexes that should **disappear** with age (normal ranges listed below):
    - Birth to 3–6 months: Moro reflex (must resolve for the child to roll)
    - Birth to 4–9 months: asymmetric tonic neck reflex
      - With patient supine, rotate head
      - Examiner should see extension of limb on the chin side and flexion on occiput side
    - Birth to 9 months: palmar and plantar grasp
  - Reflexes that should **appear** with age (normal range of appearance listed below):
    - 4–5 months: anterior propping (child should extend arms when sitting)
    - 6–12 months: parachute (may not be complete until 11–12 mo)
      - Suspend child horizontally about the waist, face down, then project the infant suddenly toward the floor or table
      - The infant should extend arms and spread fingers
    - 6–7 months: lateral propping (child should extend arms to the side if falling from sitting)
- Sensory
  - Most subjective part of a neurological examination
  - Child should distinguish light touch, pinprick (eg, from a safety pin), temperature, and vibration
    - Anterior cord: pain, fine touch, and temperature can be distinguished when intact
    - Posterior cord: proprioception, vibration, and 2-point discrimination can be determined when intact
  - If concerned, check for levels on the torso and extremities

- If a deficit is noted, complete a further examination to identify distribution and likely location
  - Check dermatomes, nerve roots, and peripheral nerve distribution
  - Cortical: all modalities; remember the sensory homunculus
  - Higher cortical testing: Rhomberg's test
    - Patient stands with feet together and arms fully extended, palms up, eyes closed
    - Observe the patient for swaying or falling to one side or another; note if patient always falls to the same side on repeated testing

- Gait
  - Native gait: should be appropriate for the patient's age
    - Note symmetry and amount of arm swing, stability, and toe walking (if any)
    - Note the patient's ability to walk in a straight line
    - Observe for ataxia, painful gait, and other abnormal gait patterns
  - Have patients walk on their toes, heels, in tandem (toe–heel), and run
    - Observe arm swing and gait abnormalities
    - These exercises are considered "stressed gaits"
    - Observe for upper-extremity posturing, including flexion at the elbow or wrist, cortical thumbing, fisting, internal rotation of the arm, or other abnormal posturing

Evaluating for Hypotonia in Infants

- Evaluation (Table 31-1)
  - Passively flap infant's hands and feet and note tone
  - Scarf sign
    - Place the infant in a semireclined position
    - Grasping the infant's hand, pull the infant's arm across the chest toward the opposite shoulder
    - If the elbow passes the midline, the patient is abnormally hypotonic
  - Vertical suspension
    - Support the patient in the axilla
    - Patient should not slip through the examiner's hand

Table 31-1. Distinguishing Examination Features of the Hypotonic Infant

Source	Tone	Strength	Sensation	Atrophy	Fasciculations	Deep Tendon Reflexes
Brain	Decreased truncal	Normal	Normal	Disuse	No	Normal or increased
Spinal cord	Above lesion: normal Below: decreased	Above lesion: normal Below: decreased	Above lesion: normal Below: decreased	Below	Occasionally below	Above lesion: normal Below: increased
Anterior horn cell	Decreased	Decreased	Normal	Occasionally	Marked	None
Nerve	Decreased	Decreased distal	Decreased distal	Present	Present	Decreased
Neuromuscular junction	Slight decrease	Fluctuates; worse with activity	Normal	None	None	Occasionally decreased
Muscle	Decreased	Decreased	Normal	None	None	Decreased over time
Ligament	Decreased	Normal	Normal	None	None	Normal

and should be able to maintain a sitting position
  - Horizontal suspension
    - Suspend the prone infant above a table while supporting the abdomen
    - Infant should lift head and bottom, arching back
    - If hypotonic, the infant's head and legs will hang down
  - Traction response
    - Place thumbs in the infant's palms and fingers around the infant's wrists
    - Gently pull the infant from the supine position
    - Infant should flex at the elbows, and head should rise and be maintained briefly in the axis of the trunk, even in full-term newborns
    - The time in which the head is aligned with the body should increase as the infant gets older
- History
  - Prenatal
    - Prenatal infections
    - Drugs
    - Fetal movement
  - Birth
    - Full-term or premature
    - Vaginal or cesarean-section birth
    - Breech or regular delivery
    - Complications
    - Newborn nursery or newborn intensive care unit
    - Time until discharged home
  - Feeding well or poorly
  - History of consuming honey or corn syrup
  - Consumption or construction in child's neighborhood
  - Static versus acute onset
  - Any developmental regression (or loss of milestones or abilities they previously could perform)
  - Family history
    - Muscular dystrophy
    - Myotonic dystrophy
    - Neuromuscular disorders
    - Sudden infant death syndrome
    - Consanguinity

        - Relatives requiring assistance to walk
- Localization
    - Brain
        - Causes include: hypoxic ischemic encephalopathy; stroke; prematurity; metabolic, chromosomal, and peroxisomal disorders; malformations; and delayed myelination
        - Head and trunk will be floppy; arms and legs may have increased tone
        - Sophisticated workup includes magnetic resonance imaging of the brain, chromosomal analysis, fluorescence in situ hybridization (FISH), positive or negative metabolic laboratory examinations, and specialist consultation, if available
    - Spinal cord
        - Lesions here are causes of hypotonia
        - Trauma, myelomeningocele, and tumor may be causes
        - Perform magnetic resonance imaging if possible
    - Anterior horn cell
        - Spinal muscular atrophy can cause hypotonia
            - Werdnig-Hoffmann disease (early)/type II
            - Kugelberg-Welander disease
        - Polio
        - Genetic disorders (sensory motor neuropathy) can also cause hypotonia
    - Nerve
        - Also a rare cause of hypotonia
        - Other causes include polyneuropathies, Guillain-Barré syndrome, and hereditary neuropathy
        - Test using electromyogram or nerve conduction, if available
    - Neuromuscular junction
        - Causes include botulism and myasthenia
        - Test a stool sample for botulinum toxin or myasthenia acetylcholine receptor antibodies, respectively, for confirmation (these tests are unlikely to be available outside large medical centers)
    - Muscle
        - Causes can include muscular dystrophy (progressive),

myopathy, human immunodeficiency virus, influenza, medications (eg, azidothymidine, steroids), inflammatory disorders (eg, dermatomyositis), Addison disease, Cushing disease, hypothyroidism, and hypophosphatemia
    - Metabolic causes include glycogen storage, Pompe's disease, and mitochondrial disease
    - Test using creatine kinase, biopsy, erythrocyte sedimentation rate, complete blood count, thyroid function test, lactate or pyruvate test, or blood chemistry test as directed by examination and other findings
  - Ligament
    - Causes may include Marfan syndrome, Ehlers-Danlos syndrome, hypermobility syndrome
    - Genetics can also cause ligament disorders that lead to hypotonia

Headaches in Children

- Acute headache
  - Defined as a single event without history of previous similar events
  - Can be generalized or localized
  - Can occur with or without neurological symptoms and signs
  - Types
    - Acute generalized: causes include fever, systemic infection, central nervous system (CNS) infection, toxins (eg, carbon monoxide, amphetamines), postictal state, hypertension, shunt malfunction, hypoxia, hypoglycemia, lumbar puncture, trauma, CNS hemorrhage, embolus, exertion, or electrolyte imbalance
    - Focal acute: causes include trauma, sinusitis, otitis, pharyngitis, Chiari malformation, glaucoma and other ocular disorders, temporomandibular joint disorder, dental disorder, or occipital neuralgia
- Acute recurrent
  - Periodic headaches that are separated by pain-free intervals
  - When associated with nausea, vomiting, and photophobia,

these headaches are usually migraines
    - Differential includes migraine; hypertension; vasculitis; substance abuse; shunt malfunction; atrioventricular malformation; mitochondrial myopathy, encephalopathy, lactic acidosis, and stroke-like episodes; postictal state; hypoglycemia; exertion; colloid cyst of the third ventricle; and dialysis
- Chronic progressive
    - Worsen in frequency and severity over time
    - May progress rapidly or slowly
    - May be accompanied by symptoms and signs of increased intracranial pressure (ICP) or progressive neurological disease
    - Neurological examination is frequently abnormal
    - Organic process/abnormality is usually present
    - Further investigation is usually warranted
    - Differential includes hydrocephalus, subdural hematoma, neoplasm, abscess, Dandy-Walker complex, Chiari malformation, subdural empyema, and pseudotumor cerebri
- Chronic nonprogressive
    - Chronic daily migraines (transformed migraines) begin with episodic migraines and a number of years later (depending on medication overuse and headache frequency) headaches evolve into daily vascular headaches
    - Chronic tension-type headaches may occur several times a week, more than 15 days a month, or may be constant
        - Usually not associated with symptoms of increased ICP or progressive neurological disease
        - Neurological examination is normal
        - Often related to school, stress, family dysfunction, or medication overuse

Evaluation of Children and Adolescents with Recurrent Headaches

- Diagnosed on a clinical basis rather than by testing
    - Diagnostic studies are not indicated when the clinical history shows no associated risk factors and the child's examination is normal
    - Lumbar puncture and electroencephalogram (EEG) are

not recommended unless history is concerning for CNS infection or seizure
    - EEG helps rule out benign occipital epilepsy, which presents with visual disturbances and can be associated with migraines
- Neuroimaging
    - Not recommended on a routine basis
    - Consider in children with abnormal neurological examinations
    - Concerning signs include focal findings, signs of increased ICP, significant alteration of consciousness, and coexistence of seizures
    - Consider in children with history of recent onset of severe headaches, change in type of headaches, or if associated features suggest neurological dysfunction
- Migraine presentation and management in children and adolescents are similar to adult presentation and management
    - Be cautious with drug dosing and aware that some medications are not approved for use in young children